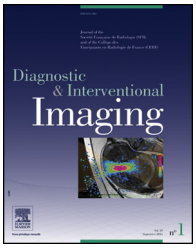
Diagnostic and Interventional Imaging (2014) **95**, 323—324



LETTER / *Musculoskeletal imaging*

Systemic lupus erythematosus with polyarthralgias due to myelofibrosis in a 49-year-old female

Keywords: Lupus; Polyarthralgia; Myelofibrosis; IRM; Pancytopenia

Systemic lupus erythematosus (SLE) is an autoimmune disease with multiple complications in multiple organs.

Our article is about osseous aspect of SLE myelofibrosis, which is an infrequent but typical complication.

Observation

A 49-year-old woman suffering from systemic lupus erythematosus for the past 15 years, treated by corticotherapy, complained about chronic polyarthralgias during 6 months. Hematological state only shows pancytopenia without other abnormalities. Initial radiological evaluation included shoulder and knee standard radiographs. Shoulder and knee MRI were also carried out to complete the investigation.

Standard radiographs were taken on heterogeneous spongious bones, including epiphyseal, metaphyseal, as well as diaphysal areas on humeral, acromion, clavicle, patellar, tibial, and femoral bones. The lesions are mostly lytic, multilocular (Fig. 1 a and b) with conservation (Fig. 1c) of some aspect of trabecular bones or sclerosis, which appear laminated. Endosteal lysis below 50% is present (Fig. 1 a). There is neither periosteal apposition nor lesions in soft tissue.

The MRI confirms the presence of several heterogeneous epiphyseal, metaphyseal, and diaphysal lesions presenting hyposignal on T1-weighted images, as well as rounded, well-defined cystic lesions with hypersignal on T2-Fat Sat weighted images (Fig. 2a—c).Their fluid level is consistent with bleeding lesions. Furthermore, there is peripheric enhancement on T1 post-gadolinium weighted images (Fig. 2a—c). There is no intra-articular effusion, and the soft tissue is normal.

These multi-focal lesions in an SLE patient with isolated pancytopenia initially resemble multi-focal bone myelofibrosis.

Renal and autoimmune laboratory tests are normal. Blood smear shows dacryocytes, along with some myelocytes and metamyelocytes. Osteomedullar biopsy shows no presence of malignant cells (blast cells) with a decrease of global cells and an increase of megakaryocytes. Argentic coloration shows an increase of reticulin fibers, consistent with medullar fibrosis. Therefore, the final diagnosis is myelofibrosis.

Discussion

The main cause of myelofibrosis is the imbalance between collagen deposition and collagen destruction by collagenase [1]. In SLE patients, platelet-derived growth factor (PDGF) and transforming growth factor-β (TGF-β) stimulate collagen destruction are increased by autoantibodies

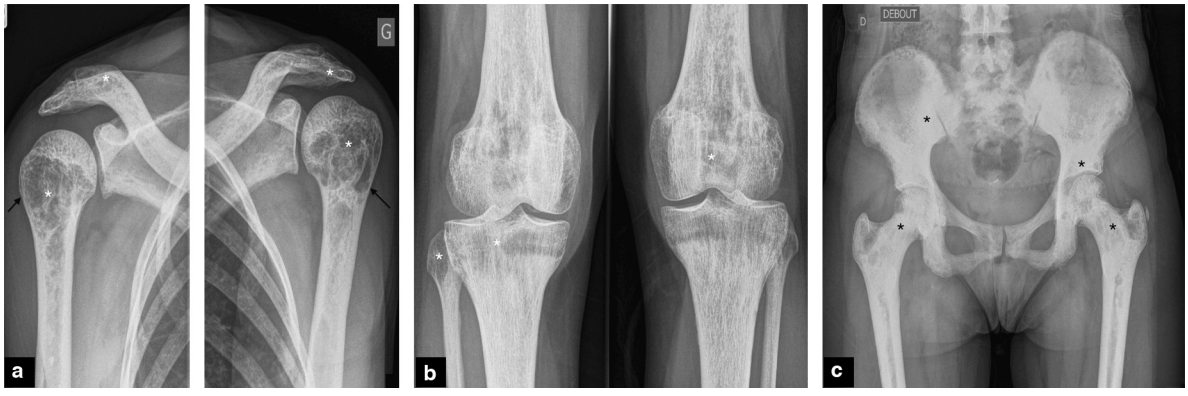


Figure 1. a: radiographs showing multilocular lytic lesions of the left and right humerus, b: radiographs showing multilocular lytic lesions of the knees; c: radiographs showing sclerosis of the pelvic bone. The white asterisks show the location of the multilocular lytic lesions. The black asterisks indicate the sclerosis of the bone. The arrows show the endosteal lysis.

2211-5684/$ — see front matter
http://dx.doi.org/10.1016/j.diii.2013.12.004

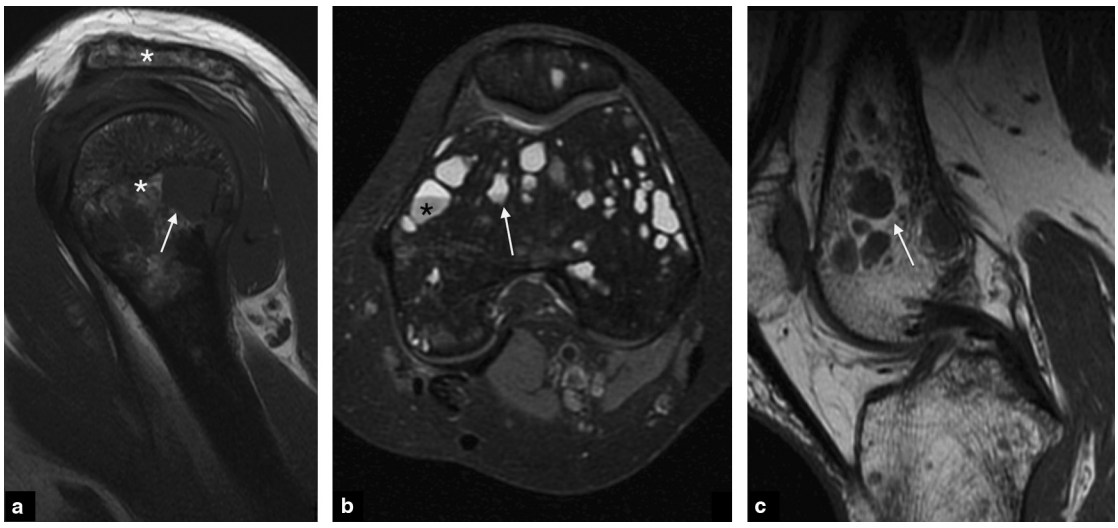


Figure 2. a: T1 post-gadolinium weighted images showing multilocular lytic lesions of the humerus; b: T2-Fat Sat weighted images showing multilocular lytic lesions of the knee; c: T1-weighted images showing multilocular lytic lesions of the knee. The white arrows and the white asterisks show the location of the cystic lesions.

fixed on megakaryocytes. SLE patients with myelofibrosis show increased procollagen 1 and 2 in serum, evidence of procollagen synthesis by medullar fibroblasts stimulated by PDGF and TGF-β. Corticotherapy has an immunosuppressive effect, which classically decreases the number of autoantibodies fixed on megakaryocytes, and thus, decreases fibrosis [2,3]. In this case, despite corticotherapy long-term treatment, sclerosis is really present in radiographs, representing the only atypical aspect of the myelofibrosis under treatment.

Hematologic disorders affect 85% of SLE patients. However, myelofibrosis is a rare case of SLE pancytopenia [3—6]. In most cases, chronic inflammatory autoimmune anaemia (70%), thrombopenia by anti-phospholipid syndrome, or hypersplenism is observed [7,8].

The diagnosis was confirmed by the pathologist from a bone biopsy.

Conclusion

Myelofibrosis is an uncommon complication of SLE, but the typical radiologic aspect needs to be known to avoid misdiagnosis.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References

[1] Kiss E, Gál I, Simkovics E, Kiss A, Bányai A, Szakáll S, et al. Myelofibrosis in systemic lupus erythematosus. Leuk Lymphoma 2000;39:661—5.
[2] Paquette RL, Meshkinpour A, Rosen PJ. Autoimmune myelofibrosis: a steroid-responsive cause of bone marrow fibrosis associated with systemic lupus erythematosus. Medicine 1994;73:145—52.
[3] Aziz AR, Mohammadian Y, Ruby C, Momin Z, Kumar A, Griciene P, et al. Systemic lupus erythematosus presenting with pancytopenia due to bone marrow myelofibrosis in a 22-year-old male. Clin Adv Hematol Oncol 2004;2(7):467—9.
[4] Feng CS, Ng MH, Szeto RS, Li EK. Bone marrow findings in lupus patients with pancytopenia. Pathology 1991;23:5—7.
[5] Daly HM, Scott GL. Myelofibrosis as a cause of pancytopenia in systemic lupus erythematosus. J Clin Pathol 1983;36:1219—22.
[6] Pereira RM, Velloso ER, Menezes Y, Gualandro S, Vassalo J, Yoshinari NH. Bone marrow findings in systemic lupus erythematosus patients with peripheral cytopenias. Clin Rheumatol 1998;17:219—22.
[7] Pavithran K, Raji NL, Thomas M. Aplastic anemia complicating systemic lupus erythematosus: report of a case and review of the literature. Rhumatology Int 2002;22:253—5.
[8] Boumpas DT, Fessler BJ, Austin 3rd HA, Balow JE, Klippel JH, et al. Systemic lupus erythematosus: emerging concepts; Part 1. Ann Inter Med 1995;122:940—50.

A. Fechner[a,b], V. Hummel[a], M. Moustarhfir[a,b], J. Manelfe[a,b], E. Schouman-Claeys[a,b], B. Dallaudière[a,b,c,*]
[a] *Department of radiology, Hôpital universitaire Bichat, 46, rue Henri-Huchard, 75018 Paris, France*
[b] *Université Paris-Diderot, 5, rue Thomas-Mann, 75013 Paris, France*
[c] *Inserm U698, Hôpital universitaire Bichat, 46, rue Henri-Huchard, 75018 Paris, France*

* Corresponding author. Department of radiology, Hôpital universitaire Bichat, 46, rue Henri-Huchard, 75018 Paris, France.
E-mail address: benjamin.dallaudiere@gmail.com (B. Dallaudière)